Congenital Heart Disease in Cyanotic Children

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• *Cyanosis is often the only apparent symptom of congenital heart disease for which a child is brought to a physician. Some of the more common anomalies can be diagnosed from this and other symptoms by a general practitioner. Squatting after exertion is a sign of tetralogy of Fallot; severe disability with relatively mild cyanosis may indicate pure pulmonary stenosis. A brisk, short, rasping systolic murmur is characteristic of these conditions and of tricuspid atresia.*

Tetralogy of Fallot is further symptomatized by a boot-shaped heart, not greatly enlarged, and right axis deviation on electrocardiograms. Typically the lung fields are clear. The author's treatment of choice is aortic-pulmonary or subclavian-pulmonary anastomosis as indicated, preferably done after the child is three years old if the condition is not so severe as to require earlier operation.

Pure pulmonary stenosis, which in some cases cannot be distinguished from tetralogy of Fallot except by cardiac catheterization and angiocardiography, may in more typical cases be diagnosed by convexity rather than concavity in the pulmonary segment and by differences in electrocardiograms. An expanding valvulotome is used to open the stenosed pulmonary valve, which is then dilated.

A systolic murmur, a round heart and left axis deviation are usually found in tricuspid atresia. Shunt operations performed for relief of this condition may lead to later heart failure because of the devious rerouting of blood through the heart.

The operations here outlined and others are statistically evaluated.

AS A RESULT of newspaper publicity during recent years almost every mother knows something about "blue babies." As it is not difficult to detect cyanosis even when confined to the lips and fingers, a child may be taken to a physician because of this symptom alone. In some cases the child is cyanotic from birth; in others after a few months of normal life, cyanosis begins to appear on crying. The cyanosis is deep and constant in some cases, while in others it appears only after exertion.

If it is reported that the child squats after unusual exertion, tetralogy of Fallot should be suspected. On the other hand, if cyanosis is relatively mild but the child is greatly incapacitated, pulmonary stenosis with intact interventricular septum—so-called pure or isolated pulmonary stenosis—may be present.

A general physical examination of weight, height, nutrition and disposition gives an immediate impression of the severity of the disease, but attention should be directed primarily to the heart sounds. Usually in a patient with tetralogy of Fallot or pure pulmonary stenosis or tricuspid atresia, a rather brisk, short, rasping systolic murmur may be heard over the precordium and to the left of the sternum at the second or third interspace. If the murmur is heard in diastole it is not typical of those conditions and probably indicates a diagnosis beyond the ability of most general practitioners. If no murmur can be heard and the cyanosis is deep the child very likely has atresia, not stenosis of the pulmonary artery, and is surely a poor operative risk.

The number of erythrocytes in the blood of cyanotic children varies from 5 to 11 million per cubic millimeter and the hemoglobin content is correspondingly high.

Roentgen examination and electrocardiographic studies are most important:

1. A child with tetralogy of Fallot usually has a boot-shaped heart because of a concavity at the base of the heart on the left in the region of the pulmonary segment, but the heart may be normal in shape, especially if the symptoms are mild. Rarely in this disease is the heart much enlarged. If it is, some complicating condition or another cardiac anomaly may be present, and shunt operation should be avoided if possible because children with enlargement of the heart do not tolerate this operation well. Typically in tetralogy of Fallot an electrocardiogram shows deviation of the axis to the right, manifesting right heart strain, while the lung fields appear clear on fluoroscopy as well as in x-ray films.

2. Pure pulmonary stenosis is symptomatically similar to tetralogy of Fallot althought the patho-

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logic changes and the treatment are quite different. The child is usually more incapacitated than would be expected from the degree of cyanosis. The pathologic change is practically limited to the valve of the pulmonary artery, where the cusps are partially fused and thereby form a barrier to the flow of blood. In tetralogy of Fallot there is a septal defect which allows escape of blood to the left ventricle, but in patients with pure pulmonary stenosis the septum is intact and the only escape for the blood dammed by the stenotic pulmonary valve is through a patent foramen ovale. Consequently x-ray films of the heart usually show enlargement with *convexity* in the region of the pulmonary segment. The electrocardiogram indicates marked right heart strain and a high P wave in lead II. As in tetralogy of Fallot, the lung fields usually are clear.

3. Tricuspid atresia is not very common. The history and general symptoms mimic those of the tetralogy of Fallot. Because the tricuspid valve is occluded, blood returning from the body must go from the right auricle through a patent foramen ovale to the left auricle and thence to the left ventricle. From there some of the blood is pumped into the aorta and some of it goes through a ventricular septal defect into the pulmonary artery. It is obvious why children with this disease are greatly incapacitated and are usually brought to the physician during infancy. The heart is round and the right ventricle diminutive, not hypertrophic as in tetralogy of Fallot; consequently the electrocardiogram indicates *left* heart strain. A cyanotic infant with a systolic murmur, a round heart, clear lung fields and deviation of the axis to the *left* probably has tricuspid atresia.

The above are the cardinal points in the diagnosis of the more common types of cyanotic heart disease which are amenable to operation. Space does not permit discussion of transposition of the great vessels, persistent truncus arteriosus and a number of less common anomalies seen in infants and to date not successfully treated surgically. Detailed information on diagnosis is available in Dr. Helen Taussig's "Congenital Malformations of the Heart."

When should cardiac catheterization be done and when should angiocardiograms be made? Only when a definitive diagnosis cannot be made without them. The typical case of tetralogy of Fallot or tricuspid atresia requires neither study. Differentiation of somewhat atypical tetralogy of Fallot from pure pulmonary stenosis usually requires both, and even with this aid a positive diagnosis cannot always be made. Differentiation of transposition of the great vessels from tetralogy of Fallot likewise occasionally demands these methods. Both procedures are valuable but not by any means conclusive. Nothing take the place of the clinical acumen of a well trained cardiologist. It must be remembered that angiocardiograms show only fleeting shadows, not always true images. Furthermore, angiocardiography is not without danger.

Surgical relief of cyanosis due to congenital heart disease was brilliantly introduced by Blalock and is now accepted. At present surgical procedures are used in three diseases to increase the flow of blood to the lungs.

In tetralogy of Fallot, if the general condition of the child is such that it can thrive and not be in danger of cerebral accidents it is advisable not to operate before the child is about three years old, as later discussion of mortality rates will explain. However, if it is obvious that the child cannot live without relief, the operation should be done regardless of age, and should be done regardless of the child's condition if it can be demonstrated that the cause is diminished flow of blood to the lungs.

The operator will probably use the surgical technique with which he is most familiar. The author opens the chest through the left fourth interspace in all cases except for infants below one year of age whose aortic arch is on the right. If the child has a left aortic arch an aortic-pulmonary anastomosis is done. If the arch curves to the right, the innominate artery being on the left, a subclavian-pulmonary anastomosis is done. In infants below one year of age the subclavian artery is too small for a suitable anastomosis and therefore—if the arch curves to the right—aortic-pulmonary anastomosis is done on the right side. An aortic-pulmonary anastomosis on the left side is technically simple, whereas on the right side it is difficult because the pulmonary artery is short and runs at almost a right angle with the aorta.

Brock of England has advised a transventricular approach to the stenosis and with a rongeur forceps blindly nips out bits of tissue in the obstructing infundibular region. Theoretically it is logical to attack the stenosis, the most significant pathological condition. Practically it is very difficult to know what one is doing when blindly and hastily cutting tissue out of the inside of the heart. The far greater mortality accompanying intracardiac surgery as compared with one of the shunt operations makes the latter procedures very emphatically preferable. When it will become possible by means of artificial heart and lungs safely to deflect the flow of blood from the heart, then intracardiac surgery for the treatment of tetralogy of Fallot will undoubtedly be the procedure of choice.

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TABLE 1.—*Mortality Following Operation for Congenital Heart Disease (aortic-pulmonary or subclavian-pulmonary anastomosis)*

Age at Operation	Number	Deaths	Percentage
TETRALOGY OF FALLOT			
2 weeks to 3 years	133	20	16.0
3 to 16 years	161	4	2.5
Total	294	24	8.0
PURE PULMONARY STENOSIS			
23 days to 11 years	19	1	5.3
TRICUSPID ATRESIA			
11* days to 3 years	12	6	50.0
3 to 11 years	5	0	0.0
Total	17	6	35.0
EXPLORATORY OPERATIONS	25	11	44.0

* Five of the six deaths were in infants below six months of age.

strain on the heart may cause heart failure. Some surgeons have attempted to improve the flow of blood by enlarging the foramen ovale at the time the shunt operation is done. The added risk of this procedure offsets the possible postoperative improvement.

Pure pulmonary stenosis is treated successfully surgically only by the method advised by Sellors and Brock of England. An expanding valvulotome (devised by the author) is thrust through the right ventricle and guided through the stenotic pulmonary valve. Immediately there is marked improvement of the patient's condition. The valvulotome is then withdrawn and a dilator introduced to open the valve to the diameter of the pulmonary artery.

A shunt operation is definitely contraindicated in pure pulmonary stenosis because of the added load placed upon the heart. For example, tetralogy of Fallot was diagnosed in an 18-month-old child and an aortic-pulmonary anastomosis was done. A year later the child's heart was enormous. The error in diagnosis was recognized. A second operation was done in which the aortic-pulmonary anastomosis was taken down and the stenotic valve incised by introducing the valvulotome through the wall of the main pulmonary artery and directing it throught the stenotic valve. The child made an uneventful recovery and the heart has decreased in size.

As to transposition of the great vessels, the mortality is so high and the results are so poor that any operation devised to date hardly seems worthwhile. Much thought has been given this problem but no answer has been found, primarily because the coronary arteries arise from the pulmonary artery and even if the vessels could be reversed the coronary arteries cannot be moved to the aorta.

Because of various methods of grouping cases it is impossible to compare one set of mortality statistics with another. It has been the author's policy to divide the patients into two groups—those in the first three years of life and those above three years (Table 1). A few significant facts at once present themselves. The outlook for patients with tetralogy of Fallot operated upon after three years of age is good, with a mortality of only 2.5 per cent, whereas the mortality in children below three years of age is 16 per cent. Obviously operation should be postponed until the patient is more than three years old unless the disease is so severe as to outweigh the factor of higher mortality in younger children.

In previous reports on mortality, cases of tricuspid atresia were included in the group of tetralogy of Fallot. In this presentation they are considered separately and the operative mortality is 35 per cent.

If, for some reason such as absence or small diameter of the pulmonary artery, no blood can be shunted to the lungs, the exploratory procedure is followed by a very high mortality, 44 per cent.

Although a number of the patients with pure pulmonary stenosis were critically ill and seemed on the verge of heart failure, the mortality has not been high. In no other congenital heart disease are the results more spectacular.

Results following operation in general have been good. In answer to the question, not infrequently put, "Is the effort worth while?" the answer is definitely, yes. Not all patients do well; some have extensive postoperative enlargement of the heart and some die, but the large majority are tremendously improved. They can go to school, ride bicycles, and run and play with other children. To be sure, the heart muscle has been subjected to an added strain by the fashioning of an artificial ductus, but it seems to stand the strain very well. Life expectancy is far from normal, because the basic dysfunction has not been corrected and, what is more, a burden has been added; but follow-up studies on children who were operated upon in 1946 and 1947 suggest that cardiac enlargement is not progressive.

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